

# A Case of Acral Persistent Papular Mucinosis

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Acral persistent papular mucinosis has been thought to be a distinct form of cutaneous mucinosis not associated with systemic diseases. It was recently classified as one of five subtypes of localized lichen mixedematosus. A 64-year-old man presented with a 15-year history of flesh-colored to translucent papules and a few round hypopigmented patches on his wrists, back of the hands, and distal forearms. Biopsy from the papular lesions revealed focal mucin accumulation in the upper reticular dermis. The clinical and histopathological features were consistent with acral persistent papular mucinosis. (*Ann Dermatol* 15(1) 8~11, 2003).

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Cutaneous mucinoses are a heterogeneous group of disorders characterized by an abnormal deposition of mucin in the dermis. Acral persistent papular mucinosis (APPM) is an unusual form of cutaneous mucinosis characterized by multiple discrete papules located symmetrically on the back of the hands, wrists, and distal forearms, focal mucin deposits in the upper dermis, slow progression over several years, and an absence of systemic involvement<sup>1</sup>. In addition to the first case of APPM reported by Kim et al<sup>2</sup> in 1993, two more cases have been reported by Lee<sup>3</sup> in 2000. We present a typical case of APPM with a review of literature.

## CASE REPORT

A 64-year-old man presented with a 15-year history of asymptomatic cutaneous lesions symmetrically located on his wrists, back of the hands, and distal forearms (Fig. 1). Physical examination revealed flesh-colored or translucent papules and a

few well-demarcated, round hypopigmented patches among them. Individual papules were dome-shaped, firm, and smooth on the surface. They persisted since they developed 15 years ago. He had subtotal gastrectomy due to severe gastric ulcer 15 years ago. Otherwise, his past history and family history were nonspecific. His complete blood count revealed mild anemia with hemoglobin 11.9g/dl, but his ferritin, TIBC, PB smear were all in normal range. Urinalysis, liver function test, thyroid function test were shown to be normal or within normal limit. The biopsy was done at the papule and the hypopigmented patch, respectively. Hematoxylin-and-eosin-stained sections of a papule (Fig. 2) demonstrated the presence of a pathologic zone in the upper part of the reticular dermis. The collagen fiber bundles were separated by empty-looking to slightly basophilic areas. Those empty-looking spaces and slightly basophilic areas were stained positively with alcian-blue at pH 2.5 (Fig. 3a) and negatively with alcian-blue at pH 1.0. They showed metachromasia with toluidine blue at pH 7.0 (Fig. 3b) and pH 4.0 but no metachromasia at pH 1.0, suggesting the presence of hyaluronic acid. The sections of the biopsy from the round hypopigmented patch did not show any evidence of mucin deposit in the dermis. It was confirmed to be an idiopathic guttate hypomelanosis with markedly reduced melanin pigment. No further treatment has been done to him.

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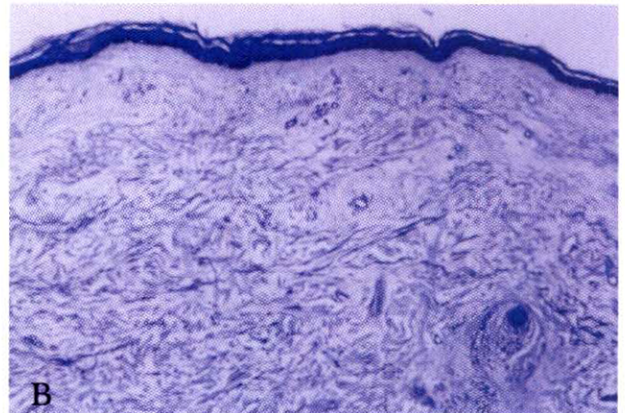
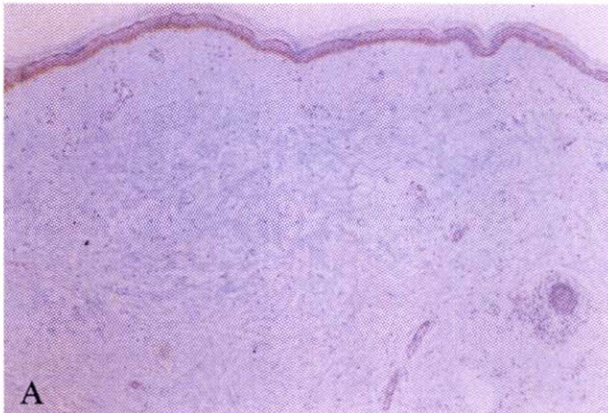
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**Fig. 1.** Asymptomatic flesh-colored or translucent papules and a few hypopigmented, well demarcated round patches among them symmetrically located on patient's distal forearm



**Fig. 2.** Biopsy from the papule demonstrates the separation of collagen fiber bundles by empty-looking areas in the upper dermis. (H&E stain,  $\times 40$ ). The hypopigmented patches among the papules were found out to be idiopathic guttate hypomelanosis.



**Fig. 3.** Those empty-looking spaces between collagen fibers are stained positively with alcian-blue at pH 2.5(A). They showed metachromasia with toluidine blue at pH 7.0(B), suggesting the presence of hyaluronic acid.

## DISCUSSION

Cutaneous mucinoses are a heterogeneous group of dermatologic alterations, characterized by cutaneous deposits of acid mucopolysaccharides in a focal or diffuse form. Lichen myxedematosus is an idiopathic cutaneous mucinosis without associated thyroid disease. In 1953, Montgomery and Underwood<sup>4</sup> established a clinical classification of papular mucinosis, with four subtypes: (1) generalized lichenoid papular eruptions; (2) discrete papular form; (3) localized or generalized lichenoid plaque form; and (4) urticarial plaques and nodular eruptions. The nosology of APPM has been controver-

sial. APPM was once thought to be a form of discrete papular form of lichen myxedematosus(DPLM)<sup>5</sup>. Rongioletti et al<sup>6</sup> insisted that APPM is a new entity and they rejected the diagnosis of lichen myxedematosus. In contrast to APPM, DPLM occurs with equal frequency in men and women and the papules are located asymmetrically, sometimes coalesce in plaques, and are variably distributed over the face, trunk, axillary folds, and especially on the knees and elbows<sup>7</sup>. In addition, histologically, the deposit of mucin is diffuse in the mid and deep dermis compared to focal accumulation of mucin in APPM<sup>8</sup>. In APPM mucin spares a subepidermal grenz zone and does not spread through the adjacent



normal collagen bundles in the deep dermis<sup>7,9</sup>. Another differential point is that a remarkable fibroblast proliferation and an irregular arrangement of collagen bundles is only seen in DPLM<sup>7,9</sup>. Recently Franco *et al*<sup>10</sup> have updated the classification of papular mucinosis, lichen myxedematosus, and scleromyxedema. They divided lichen myxedematosus into two subsets: a generalized papular and sclerodermoid form(also called scleromyxedema) and a localized papular form. The criteria for localized LM is (1) generalized papular and sclerodermoid eruption (2) mucin deposition, fibroblast proliferation, and fibrosis (3) the absence of monoclonal gammopathy or thyroid disease. They defined APPM as one of the subtypes of the localized form<sup>10</sup>.

APPM is characterized by multiple, flesh-colored, ivory or translucent papules of 2-5 mm in diameter. Lesions are located symmetrically on the back of the hands and wrists, may extend to the distal third of the forearms. Usually, they are asymptomatic, but there has been a report of APPM with pruritus<sup>11</sup>. When punctured, the papules may yield a translucent, dense fluid. APPM has a benign clinical course with slow progression. It usually affects middle-aged persons with a female predominance.

Menni *et al*<sup>12</sup> reported acral persistent papular mucinosis in two sisters. A 56-year-old woman reported by Espana *et al*<sup>13</sup> to have APPM also had father suffering similar lesions in the same area with his daughter, although it was unconfirmed as they were unable to examine the patient's father. With regard to the familial incidence, environmental or genetic factors can be suggested to have a role in the pathogenesis of this disease. There also has been a report of a worsening of papular mucinosis(scleromyxedema type) after an accidental overexposure to ultraviolet B light<sup>14</sup>. 70-year-old woman presented by Abalde *et al*<sup>15</sup> also showed worsening of her APPM lesions by solar exposure. Those findings suggest that sun light may be associated with APPM.

Localized LM does not require treatment, and a wait-and-see approach is recommended. The response of APPM to topical corticosteroid has been inconsistent. There has been a report of APPM resulting in complete regression of the papular lesions over a few weeks with once-daily topical application of 0.05% clobetasol propionate<sup>16</sup>. How-

ever, the lesions slowly recurred upon withdrawal of topical steroid. Crovato *et al*<sup>17</sup> reported the lesions that were resistant to topical steroid therapy.

We present a typical example of APPM, demonstrating characteristic distribution without accompanying disease.

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